249 Liver disease in children with cystic fibrosis – The contribution of ultrasound evaluation

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Introduction: It is well recognized that liver involvement in Cystic Fibrosis (CF) patients is associated with a more adverse outcome. Although no perfect method of assessing hepatobiliary involvement in CF is currently available, abdominal ultrasound (AUS) and biochemical evaluation have been used to identify liver disease at an early stage. Considering the scarce information in Portuguese CF patients, we aimed to evaluate liver involvement in CF pediatric patients.
Methods: Observational/cross-sectional study in CF children, concerning the most recent liver evaluation (clinical, AUS and $\geqslant$1 biochemical evaluation).
Results: A total of 24 patients were included (median 10 years [1–18]; 9 males). All patients had pancreatic insufficiency; 23 had F508del mutation (17 homozygous). Ten patients (42%) had a normal evaluation (clinical, AUS, analytic) and the other patients (except 1) had AUS abnormalities.
Eight patients had hepatomegaly and from these 4 had splenomegaly.
AUS features included: heterogeneous echogenicity of hepatic parenchyma (11/24), with nodularity additionally present in 5/24, homogenous echogenicity (2/24), hepatomegaly (8/24) and esplenomegaly (4/24). Only 6 patients had AST/ALT or/and G-GT alterations.
Ursodeoxycholic acid therapy was instituted in patients with major liver involvement. In 3 patients submitted to upper endoscopy the presence of esophageal varices was confirmed and 2 are currently under variceal ligation program.
Conclusion: Despite the retrospective nature of the present data, a relatively high frequency of liver involvement in CF patients has been shown. We emphasize the diagnostic contribution of US evaluation as previously reported.

250 Portal hypertension in cystic fibrosis children – One center experience

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Background: Liver cirrhosis is the end stage of cystic fibrosis liver disease (CFLD) with an important implication on disease's outcome. Vascular complication, with occurrence of portal hypertension is an alarming stage considering the risk of variceal bleeding.
Objectives: Evaluation of severe liver disease with portal hypertension (PH) in CF patients and identification of its prevalence and risk factors.
Methods: Study evaluated prospectively, 159 patients for five years. They were routinely followed-up by clinical assessment, liver biochemical tests, ultrasound examinations (US); transient elastography, MRI, endoscopy.
Results: Fifty five patients, median age at diagnosis = 12.4 years were diagnosed with CFLD, with slight predominance of boys. Severe liver disease occurred in 11 patients (20%) of which 5 with oesophageal varices. All of them had history of meconium ileus and homozygous for F508del; 4 were boys. CFLC occurred more frequently in patients aged over 10 yrs. Pancreatic insufficiency and severe mutations was strongly associated with PH.
Conclusion: CF children older than 10 year, with history of meconium ileus, pancreatic insufficiency and severe mutation were more likely predisposed to liver cirrhosis with portal hypertension. Further studies should follow risk factor for development of CFLD and how they can be influenced, in order to prevent the development of liver cirrhosis.

251 Fecal incontinence in adult CF patients: Prevalence

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Patients with cystic fibrosis (CF) are deemed at risk of developing urinary incontinence (UI) due to repeated coughing and other factors causing increased pressure on the pelvic floor. Fecal incontinence (FI) is derived from the same mechanism but only very few data are available on its frequency. The aim was to establish the prevalence of FI in an adult population with CF.
Patients were recruited from Marseille referral center for adult CF. They were asked to fill in a self-completed anonymous questionnaire for symptom assessment of UI (Urinary Symptom Profile score) and FI (St Mark Hospital score). Clinical data and medical history were also recorded.
122 out of 190 patients completed the survey (79M/43F, mean age 30.9 yr [range 18–65], 59 [48%] were lung transplanted). Mean FEV1 was 67.3±28% predicted. Symptoms of FI were reported by 37 patients (30%, 24F [56%], 13M [16%]) and of UI by 31 patients (25.4%, 28F [65%], 3M [4%]). For 17 patients (14%, 14F [32.5%]) UI and FI were present. FI was significantly more frequent in older patients (34.27 yr vs 29.54 yr, $p = 0.03$) and in patient with associated UI ($p = 0.001$). No relationship was found between respiratory, bacterial, nutritional status, transplantation, pancreatic status, practice of physiotherapy, delivery history and FI.
In this preliminary study, FI prevalence is high in the CF adult population especially in women. It may also lead to potential deterioration of respiratory status due to reduced adherence to forced expiratory maneuvers aiming at avoiding the occurrence of FI and UI. This population needs to be aware of the problem of FI and UI as it is a treatable condition and could potentially be prevented.

252 Survey of malignancies at Scandinavian CF centers

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Introduction: Average age in the adult CF population is permanently increasing predisposing to a higher rate of malignancies. The previous survey on cases of malignancies in CF adults at Stockholm CF center revealed a growing number of malignancies with gastrointestinal (GI) cancer being predominant.
The aim of the study was to survey cases of malignancies in non-transplanted CF adults (n = ca 600) at the Scandinavian (Sweden, Denmark and Norway) CF centers.
The retrospective case analysis performed in CF adults revealed 18 cases of malignancies during the last decades. Nine patients had different forms of GI cancer (adenocarcinoma of colon, cholangiocarcinoma, adenocarcinoma of small intestine, gingival cancer as well as cancer of pancreas). Three patients had tumours of hematological origin (myeloma, acute lymphoblastic leukemia, and lymphoma). Four patients had tumours in endocrine organs (seminoma, thyriod cancer and cancer of ovarii). Two patients had cervical cancer and melanoma, respectively.
Further analysis disclosed five cases of benign tumours. In four of these pathology was found within GI tract (colon polyposis (3) and duodenal adenoma (1)). In two of the patients with colon polyposis GI cancer was later diagnosed (included above). Patient age varied between 25 and 65 years. Mean age for GI cancer was 48 years (39–65).
In children there were two cases of malignancies: Langerhans cell histiocytosis and acute lymphoblastic leukemia.
Conclusion: The data from Scandinavian CF centers demonstrates that GI cancer is the dominating form of malignancies among CF adults. However, one has to be aware of tumours of hematological and endocrine origin in CF patients.